# Outcome of Systemic-to-Pulmonary Shunts in Cyanotic Congenital Heart Disease- A 9-year Experience

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**Abstract-** The aim of this study is to evaluate early and late results of surgery in children with congenital heart disease and decreased pulmonary blood flow, who underwent a palliative systemic-to-pulmonary shunt. During the past 9 years, 157 systemic-to-pulmonary artery shunts were performed in 130 patients (69 males, 61 females) with ages from 1 day to 31 years old. They had been evaluated for their clinical effectiveness, the need for a repeat operation and the mortality and morbidity. There were 101 (77.7%) modified Blalock-Tausig (BT) shunts, 19 (14.6%) modified Waterston shunts, 8 (6.2%) Central shunts, 1 Waterston shunt, and 1 Glenn shunt created throughout the study. Tetralogy of Fallot comprised the majority of cases (113; 86.9%) while the remaining 17 (13.1%) included transposition of the great arteries with pulmonary stenosis, tricuspid atresia, pulmonary atresia, atrioventricular septal defect (canal) with pulmonary stenosis, and univentricular heart complex. Early mortality was 12.3% (16 patients). Second shunts were created in 25 (19.2%) patients. Forty patients (30.8%) have undergone subsequent intracardiac repair 37.7 ± 17.8 months after original shunt procedure. There were four (3%) late deaths. Follow-up was achieved in 105 of 114 early survivors for a period of 3 to 117 months (mean 31.7 ± 19.4 months). Modified BT shunt was performed most frequently in our service; it was associated with less closure and mortality than other types of shunt and had excellent clinical palliation and patency rates.



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**Key word:** Congenital heart disease, cardiovascular surgery, shunts

## Introduction

Although primary definitive intracardiac repair is now possible and often preferable in many patients with congenital anomalies, there is still a need for palliative systemic-to-pulmonary shunts in some cyanotic patients. For some, definitive repair may not be presently available.

For others, repair may involve placement of conduit or other prosthesis, in which case patient size is a critical factor. For still others, intracardiac correction may carry a significantly higher mortality than that for preliminary shunting and secondary repair. The modified Blalock shunt (MBTS), used by the Great Ormond Street Group (1,2), and subsequently popularized by several others (3,4), interposes a prosthetic tube between the subclavian and pulmonary arteries and its use requires less dissection and guarantees a proximal and distal anastomosis of large diameter, with the maximal amount of flow being regulated by the diameter of the patient's own uninterrupted subclavian artery, thus allowing for possible increase in shunt flow with growth. Somerville and associations (5) reported a series of patients with tetralogy malformations and Waterston shunts, who, at the time of repair, had a closure of the shunt from within the aorta. In this series of patients, there was a high mortality rate as a consequence of the residual stenosis at the level of the shunt in combination with the underdevelopment of the left pulmonary artery.

This reviews the early and late results in a series of 130 consecutive patients who underwent systemic-to-pulmonary artery anastomosis during a 9 year period. Our center is the leading cardiothoracic surgery hospital in north-western states of Iran and over 19 percent of those who underwent surgery in this center are congenital cases.

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## Patients and Methods

During the 9 year period from March 1998, through March 2005, 157 systemic-to-pulmonary artery shunts were performed in 130 patients [69 (53.1%) male, 61 (46.9%) female]. Ages of the patients ranged from one day to 31 years. Age distribution of patients underwent 130 procedures was shown in Figure 1.

Twenty (15.4%) patients were infants (<1 year, mean weight of 6.5 ± 1.3), two neonates (<1 month old), seven more than 10 years and the remaining 101 (77.7%) between 1 and 10 years old. Preoperative clinical data are depicted in Table 1.

The cardiac anatomy was tetralogy of Fallot in 113 (86.9%), the remaining 17 (13.1%) included transposition of the great arteries with pulmonary stenosis (6), tricuspid atresia (4), pulmonary atresia (2), atrioventricular septal defect (canal) with pulmonary stenosis (3), and univentricular heart complex (2). All patients had cardiac catheterization and angiocardiography prior to operation. All in-patient and out-patient records, cardiac catheterization data, and finding at subsequent intracardiac repair were reviewed. The patients were grouped according to age, type of shunt, anatomic diagnosis. Clinical and fallow-up data were then tabulated. The patients who had two or three shunt procedures are classified according to the initial procedure. There were 101 (right = 74, left = 27) modified Blalock-Tausig shunts (MBTSs), and 19 (right = 18, left = 1) modified Waterston shunts (MWSs), and 8 central shunts (CSs), and 1 Waterston shunt (WS), and 1 Glenn shunt (GS) created (Figure 2). In two patients, MBTSs was performed by cardiopulmonary bypass because of hypotension. Of 130 patients, 128 (98.5%) received prosthetic graft (polytetrafluoroethylene (PTFE) or Gore-Tex) shunts (Table 2).

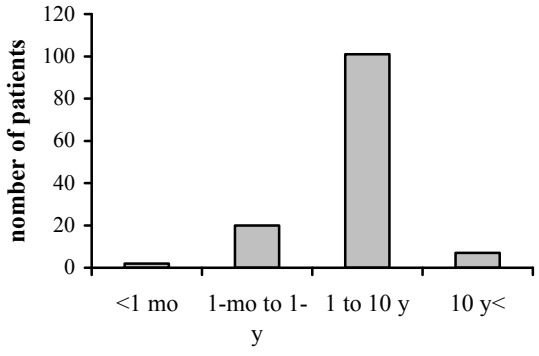


**Figure 1.** Age distribution of patients undergoing 130 shunt procedures

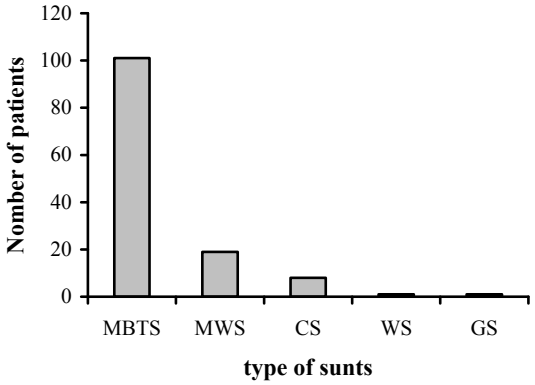


**Figure 2.** Distribution of patients according to the type of shunts applied to them. MBTS indicates Modified Blalock-Taussig shunt; MWS, Modified Waterston shunt; CS, Central shunt: WS, Waterston shunt; GS, Glenn shunt.

**Table 1.** Preoperative clinical data

| Clinical data | value | Percent |
|---|---|---|
| **Gender** | | |
| Male | 69 | 53.1 |
| Female | 61 | 46.9 |
| **Weight (kg)** | 3-51 | |
| **Symptoms** | | |
| Cyanosis | 122 | 93.8 |
| Clubbing | 46 | 35.4 |
| Dyspnea | 33 | 25.4 |
| Dyspnea on exertion | 14 | 10.7 |
| Fatigue | 27 | 20.8 |
| Palpitation | 15 | 11.5 |
| Spell | 17 | 13.1 |
| Sweating | 3 | 2.3 |
| Poor feeding | 3 | 2.3 |
| Syncope | 1 | 0.8 |
| Seizure | 3 | 2.3 |
| Cardiomegaly | 6 | 4.6 |
| Failure to thrive | 8 | 6.2 |
| Mental retardation | 3 | 2.3 |
| Trisomy 21 | 3 | 2.3 |

Among those receiving systemic-to-pulmonary shunt, a 4 mm graft in 25 ( 19.4%), a 5 mm graft was used in 74 (57.8%), and a 6 mm graft in 29 (22.7%) patients. 128 (98.5%) patients had received perioperative heparin and 116 (89.2%) aspirin, postoperatively.

**Table 2.** Distribution of shunts according to diameter of shunts

| Type of shunt* | Number(percent) | Diameter of graft ( Gore-Tex) | | |
|---|---|---|---|---|
| | | 4mm | 5mm | 6mm |
| MBTS | 101(77.7%) | **11(8.7%)** | **64(50%)** | **26(20.3%)** |
| MWS | 19(14.6%) | **8(6.3%)** | **9(7.1%)** | **2(1.6%)** |
| CS | 8(6.2%) | **6(4.7%)** | **1(0.8%)** | **1(0.8%)** |
| **total** | 128(100%) | **25(19.6%)** | **74(57.8%)** | **29(22.7%)** |

* MBTS indicates Modified Blalock Tausig Shunt; MWS, Modified Waterston Shunt; CS, Central Shunt

## Results

### Early mortality

There was no intraoperative death. In this series of 130 patients, there were sixteen deaths within 30 days of surgery or during the same hospitalization for an overall early mortality of 12.3%. There were 6 deaths among 101 patients receiving MBTS, for a mortality of 5.9%. An attempt was made to group these patients by the type of shunt procedure performed and the cause of early death (Table 3).

There were eight thrombosed shunts postoperatively that underwent subsequent shunt procedure (6) or thrombectomy (2). Four of these patients died after operation. Three patients had poor function shunt postoperatively that underwent subsequent shunt procedure and one of them died after operation. The 22 patients who were less than 1 year of age at the time of operation composed only 16.9 percent of the total number of patients but contributed 50 percent of the deaths (8 of the 16 deaths). The 101 patients who were more than 1 year and less than 10 years old composed 77.7 percent of the total series but accounted for 50 percent of the total mortality rate (8 of the 16 deaths). Of 16 early mortalities, 11 (68.8%) patients had tetralogy of Fallot, 3 (18.8%) tricuspid atresia, 1 (6.3%) pulmonary atresia, and 1 (6.3%) transposition of great artery with pulmonary stenosis.

### Early postoperative complications

Two patients had intraoperative cardiac arrest that responded to cardiopulmonary resuscitation. Postoperatiove complications included thrombosed shunt (8, 6.2%) rebleeding (7, 5.3%), acute heart failure (7, 5.3%), respiratory failure (5, 3.8%), and acute renal failure (3, 2.3%). (Table 4) One patient (MBTS) had ipsilateral pulmonary edema and dense opacification of the ipsilateral lung postoperatively that immediately returned to the operating room and narrowed the proximal anastomosis by stitch.

### Early and late shunt failures

There were eleven patients (8.5%) whom we considered early shunt failures, seven in the MBTS, 3 in the MWS, and 1 in the CS. Of eleven patients, 8 had thrombosed shunts and 3 poor function shunts. Eight thrombosed shunt underwent subsequent shunt procedures[6] and thrombectomy of the shunt (2). One of the thrombectomy shunt was poor function that second shunt was created. Four of these patients discharged from hospital.

Early shunt failure was seen in 8 tetralogy of Fallot, 1 atrioventricular defect (canal) with PS, and 2 transposition of great artery with pulmonary stenosis. There were 14 (10.8%) late shunt failures. All late shunt failures were seen in patients with tetralogy of Fallot. Eleven patients had MBTS, two MWS, and one central shunts.

**Table 3.** Causes of early mortality

| Causes of early death* | Type of shunt | | | | Total |
|---|---|---|---|---|---|
| | MBTS | MWS | WS | CS | |
| Congestive heart failure | 1 | 4 | 1 | 1 | 7(43.8%) |
| Cardiac arrest | 2 | 1 | 0 | 0 | 3(18.8%) |
| Respiratory failure | 2 | 0 | 0 | 1 | 3(18.8%) |
| Mediastinitis (septic shock) | 0 | 0 | 0 | 1 | 1(6.3%) |
| MOF | 1 | 1 | 0 | 0 | 2(12.5%) |
| Total(percent) | 6(37.5%) | 6(37.5%) | 1(6.2%) | 3(18.7%) | 16(100%) |

*MOF indicates Multiorgan Failure; MBTS, Modified Blalock-Taussig Shunt; MWS, Modified Waterston Shunt; WS, Waterston Shunt; CS, Central Shunt.

**Table 4.** Early postoperative complications

| Complications | value | Percent (%) |
|---|---|---|
| Thrombosed shunt | 8 | 6.2 |
| Bleeding (re-explore) | 7 | 5.4 |
| Acute heart failure | 7 | 5.4 |
| Respiratory failure | 5 | 3.8 |
| Acute renal failure | 3 | 2.3 |
| Sternal dehiscence | 2 | 1.5 |
| Pericardial effusion | 1 | 0.8 |
| Pleural effusion | 1 | 0.8 |
| Hepatic failure | 1 | 0.8 |
| Pneumothorax | 1 | 0.8 |
| Hematuria | 1 | 0.8 |
| Hepatitis | 1 | 0.8 |
| Ipsilateral pulmonary edema | 1 | 0.8 |

**Subsequent shunt procedures**

Second shunts were created in 25 (19.2%) patients. In the eleven early shunt failures a second shunt was created 1 to 7 (3 ± 2.86) days after original procedures because of thrombosed shunts (6) and poor functions shunts (5). In remaining fourteen late shunt failures a second shunt was created 3 to 59 (28.6 ± 14.8) months after original shunt because of poor function (10) and none function (4) shunts. The second shunt procedures were MBTSs (16), MWS (5), CS (3) and WS (1). There were 5 (20%), 16 (21%), and 4 (13%) shunt failures, in 4, 5, and 6 mm PTFE grafts consequently. Postoperative complications in these patients were bleeding (3), cardiac arrest, and respiratory failure (1). There was no death. Two patients (TOF) received three separate shunts 2 and 66 months after original shunt and one of them died because of cardiac arrest.

**Subsequent intracardiac repair**

Forty patients (30.8%) have undergone subsequent intracardiac repair 37.7 ± 17.8 months (range 10 to 85 months) after original shunt procedure. These patients had tetralogy of Fallot (39) and transposition of great artery with PS (1). These patients had received MBTS (38, 95%), and MWS (2, 5 %). There was no operative and late mortality in these patients. In all this patients, cardiac catheterization was performed before total correction and there was lack of distortion of pulmonary arterial anatomy.

**Patients without further procedures**

There are 90(69.2%) patients who remain well and free of further surgery after receiving their palliative shunts. The diagnoses and type of shunt of patients are shown in table 5.

Of 90 patients, there were four (3%) late deaths, 3 to 63 months after the original procedure, because of multiorgan failure (2), and cardiac arrest (2). Nine patients lost to follow up. Follow-up was achieved in 105 of 114 early survivors for a period of 3 to 117 months (mean; 31.7 ± 19.4 months). All surviving patients had subjective and objective improvement.

**Table 5.** Diagnosis and type of shunt of patients without procedures

| Diagnosis* | | Type of shunt | | | | | Total | |
|---|---|---|---|---|---|---|---|---|
| | | MBTS | MWS | CS | WS | GS | Number | Percent |
| TOF | | 57 | 11 | 5 | 1 | 0 | 74 | 82.2 |
| T.A | | 0 | 2 | 1 | 0 | 1 | 4 | 4.4 |
| A.V.S.D + P.S | | 2 | 1 | 0 | 0 | 0 | 2 | 3.3 |
| TGA + P.S | | 2 | 2 | 1 | 0 | 0 | 5 | 5.6 |
| P.A | | 1 | 0 | 1 | 0 | 0 | 2 | 2.2 |
| U.H.C | | 1 | 1 | 0 | 0 | 0 | 2 | 2.2 |
| Total | Number | 63 | 17 | 8 | 1 | 1 | 90 | |
| | Percent | 70 | 18.9 | 8.9 | 1.1 | 1.1 | 100 | |

*TOF indicates Tetralogy of Fallot; TA, Tricuspid Atresia; AVSD, Atrioventricular Septal Defect; PS, Pulmonary Stenosis; TGA, Transposition of Great Arteries; PA, Pulmonary Atresia; UHC, Univentricular Heart Complex; MBTS, Modified Blalock Taussig Shunt; MWS, Modified Waterston Shunt; CS, Central Shunt; WS, Waterston Shunt; GS, Glenn Shunt.

## Discussion

For patients with lesions with complex anomalies or in critically ill cyanotic newborns, definitive intracardiac repair may not be possible. For others a two-stage correction may still be preferable (6,7). It is widely accepted that the Blalock-Taussig shunt is the shunt of choice for children with pulmonary oligemia who older than 1 year (8-13). The incidence of congestive heart failure and pulmonary microvascular disease that follows the Blalock-Taussig shunt is low when compared with that of lateral shunts (8-11,13-16). Chopra and associates17 proposed that the Blalock-Taussig shunt was the shunt of choice for infants of any age with severe tetralogy of Fallot. However, the procedure requires extensive surgical dissection and sacrifice of the subclavian artery, and the maximal anastomotic diameter is limited by the size of the subclavian artery. In addition, the anastomosis tiny neonate does not keep pace with the rest of the child's growth.

Use of the modified shunt obviates some of these problems, but carries risks of it is own. It is technically simple operation, requiring less dissection and guaranteeing a wide proximal and distal anastomosis. Also, distal subclavian flow can be preserved during construction of this shunt, and it provides the potential for increased shunt flow with growth as the subclavian diameter increases. Furthermore, it allows easy creation of a shunt on the side of the aortic arch when it is necessary or preferable. Despite the fixed length of the prosthetic tube, we have not observed significant pulmonary arterial distortion with growth. Further follow-up is of course necessary, but the longer experience of other groups (1-4,18-20) shows a similar absence of distortion. At the time of subsequent surgery, the previous shunt had been simply ligated or clipped.

In most centers Blalock-Taussig shunt is the palliative procedure of choice in patients over 1 year of age (8,9,11,14). In infants under 1 year of age. However, the Waterston or Potts anastomosis has been recommended because of the small size of the subclavian artery and the previously reported poor success with the Blalock–Taussig anastomosis in this age group (8,9,11,14-16). The results of the present study indicate that the overall survival rate (94.1%) with modified Blalock shunt is better than that with the other shunt procedures, even in very young infants. The early mortality in the MBTS was 6 (5.9%) that this conclusion closely parallel those in the multicenter study by Woolf et al. (21-23). In this series, the modified Waterston shunt (MWS) had high mortality (6/19, 31.5%) versus the other study (10.7 %) (24). In the MWS, congestive heart failure was causes of early death in 4 (21%) patients which in the MBTS it was 1 (≈ 1%). On the other hand, early shunt patency for the MBTS and MWS groups were 93.1% (95/101) and 84.3% (3/19) consequently. Therefore, our results indicate that MBTS had lower early mortality and higher early shunt patency rate than MWS. Systemic heparinization (98.5%) was routinely used postoperatively. However, there was 8 thrombosed shunts after original shunts and heparinization resulted in a reoperation rate of 5.4%. There were eleven (8.5%) early and fourteen (10.8%) late shunt failures that second shunt was created 3.0 ± 2.86 days and 28.6 ± 14.8 months after original shunt, consequently. We observed no significant difference in the shunt patency rate between 4mm (80%) and 5 mm (79%) grafts for a period 3 to 117 months. However, the work of Woolf et al. (21) demonstrated a rapid decrease in patency as a result of pseudointimal proliferation after 41 months in a group of patients receiving PTFE (Polytetrafluoroethylene) shunts, and the work of de Leval et al(1), Di Benedetto et al (25), and Miyamuto et al (3) clearly suggests that this problem is less frequent with larger size grafts.

In summary, our experience suggests that systemic to pulmonary shunt (MBTS, MWS, WS, CS, etc.) can be safely and reproducibly constructed. This series shows an early mortality rate comparable to that of other studies, with excellent clinical palliation and patency rate. On the other hand, in our series, there was lack of distortion of pulmonary arterial anatomy in forty patients with total correction. Therefore, PTFE shunts are preferred for the management of severe pulmonary oligemia in infants and children.

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